CASE REPORT

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A rare case of gastrointestinal stromal tumor presenting with closed perforation of the small intestine

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Gastrointestinal stromal tumors are very rare mesenchymal tumors of the gastrointestinal tract with variable clinical presentations depending on the tumor size and anatomic site. The currently used diagnostic modalities cannot establish a preoperative diagnosis with 100% certainty. The treatment of choice includes complete surgical excision and/or chemotherapy. In this paper, we present a very rare case of gastrointestinal stromal tumor presenting with closed perforation of the small intestine. The patient was successfully managed with surgical excision of the tumor with segmental bowel resection and adjuvant chemotherapy. Although rare, it must be kept in mind that gastrointestinal stromal tumors can also present with closed bowel perforation.

Key words: Gastrointestinal stromal tumor, closed bowel perforation, mesenchymal tumor

İnce bağırsağın kapalı perforasyonu ile kendini gösteren nadir bir gastrointestinal stromal tümör olgusu

Gastrointestinal stromal tümörler gastrointestinal sistemin en sık görülen mezenkimal tümörleridir. Bu tümörler nadir görülür. Tümörün boyutu ve anatomik yerleşim yerine göre çeşitli klinikle ortaya çıkar. Şu anki tanı yöntemleri preoperatif tanıyı %100 desteklemez. Tedavi seçenekleri tümörün tamamının çıkarılmasını ve /veya kemoterapiyi içermektedir. Biz kapalı perforasyon ile kendini gösteren ince bağırsak kaynaklı dev gastrointestinal stromal tümörlü olan bir olguyu sunmaktayız. Hasta segmental ince bağırsak rezeksiyonu ile tümörün cerrahi çıkarılmasını takiben kemoterapi alarak başarılı bir şekilde tedavi edildi. Ender olmasına rağmen gastrointestinal stromal tümörlerin bu vakada görüldüğü gibi aynı zamanda kapalı perforasyonla da ortaya çıkabileceği akılda tutulmalıdır.

Anahtar kelimeler: Gastrointestinal stromal tümörler, kapalı bağırsak perforasyonu, mezenkimal tümör

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are very rare mesenchymal tumors of the GI tract with a frequency of approximately 10-20/1,000,000 population (1,2). The clinical presentation is variable and depends on the tumor size and anatomic site. The currently used diagnostic modalities include barium studies, computed tomography (CT) and angiography; however, none of these can establish a preoperative diagnosis with 100% certainty. The primary treatment of choice is complete surgical resection without extensive lymph node sampling. The prognosis of GISTs is worse in case of bowel perforation, metastasis or locoregional recurrence.

In this report, we aimed to present a rare case of a GIST presenting with closed perforation of the small intestine. To the best of our knowledge, such a case has not been reported previously in the literature.

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CASE REPORT

In December 2009, a 48-year-old male patient was admitted to Cerrahpaşa Medical University, Department of General Surgery with the complaints of vague abdominal pain lasting for one month. On physical examination, a palpable mass and tenderness were noted on the left upper and lower abdominal quadrants. Digital examination revealed hematochezia. The results of routine laboratory tests including tumor markers were within normal limits, except for low hematocrit level (23.8%) that was treated by erythrocyte suspensions; there was no abnormality in the biochemical results. CT examination with oral and intravenous (IV) contrast administration revealed an intraabdominal mass with peripheral enhancement, 21x15x11 cm in diameter, localized in the left paramedian area, with its origin from the small bowel and with extension to the left kidney anteriorly and urinary bladder superiorly. The mass was in close relation with the tail of the pancreas and the proximal part of the left ureter. Necrosis and air bubbles were noted in the central part of the mass, where oral contrast medium extravasation was also noted, suggesting the diagnosis of closed small bowel perforation (Figure 1).

Based on these clinical and radiologic findings, anemia was corrected with erythrocyte suspensions, and surgery was planned. Following a median laparotomy, a tumoral mass measuring 20 cm in diameter extending from the anterior abdominal wall to the left retroperitoneal space was seen (Figure 2). On further exploration, the mass was found to originate from the proximal jejunum, and a closed small bowel perforation was determined in this intestinal segment. No metastasis was de-

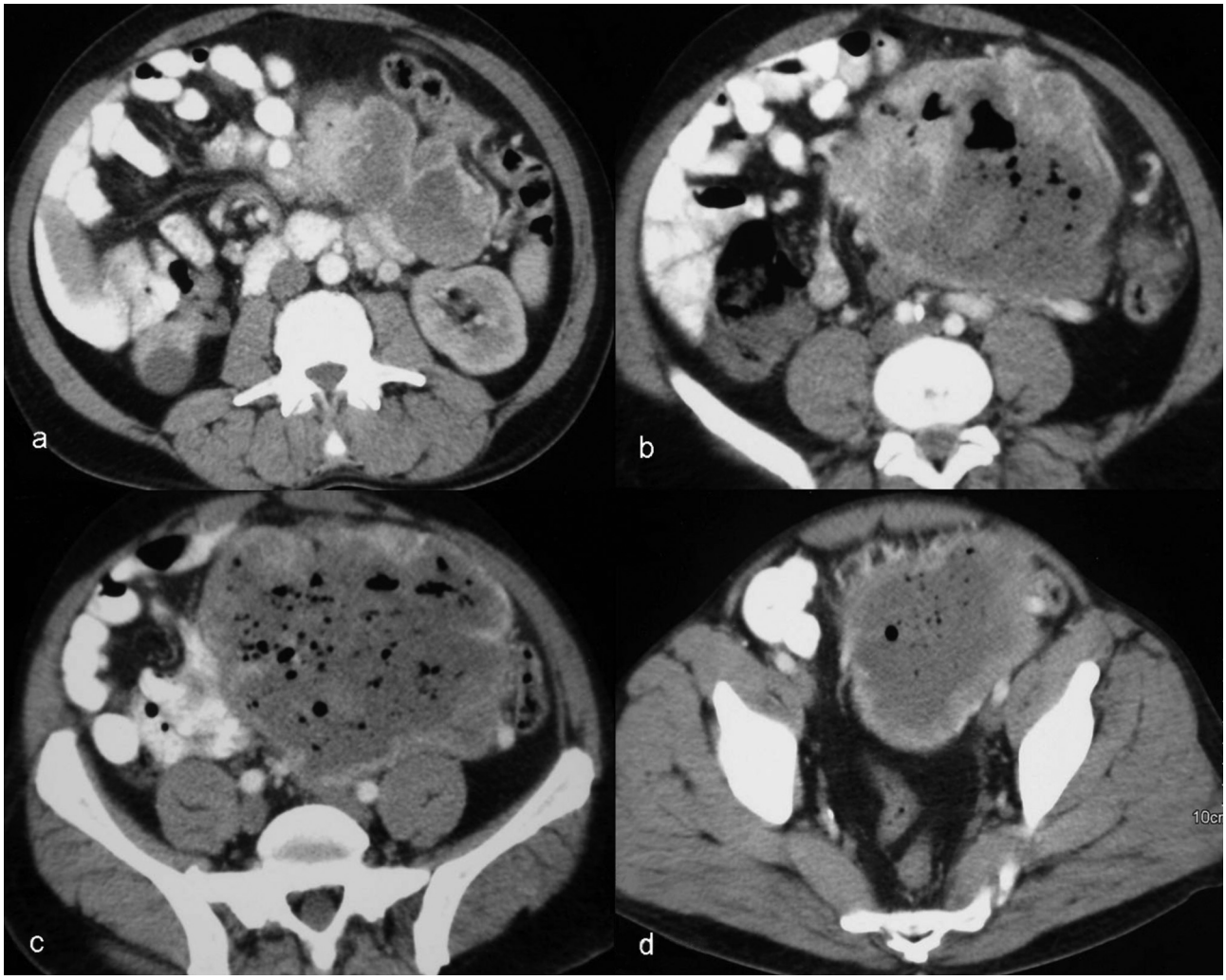

Figure 1. Axial CT scans show **(a)** the upper margin of the tumoral mass, **(b)** the presence of extravazated oral contrast medium (arrow), **(c)** multiple air bubbles within the necrotic mass and **(d)** the lower margin of the mass.

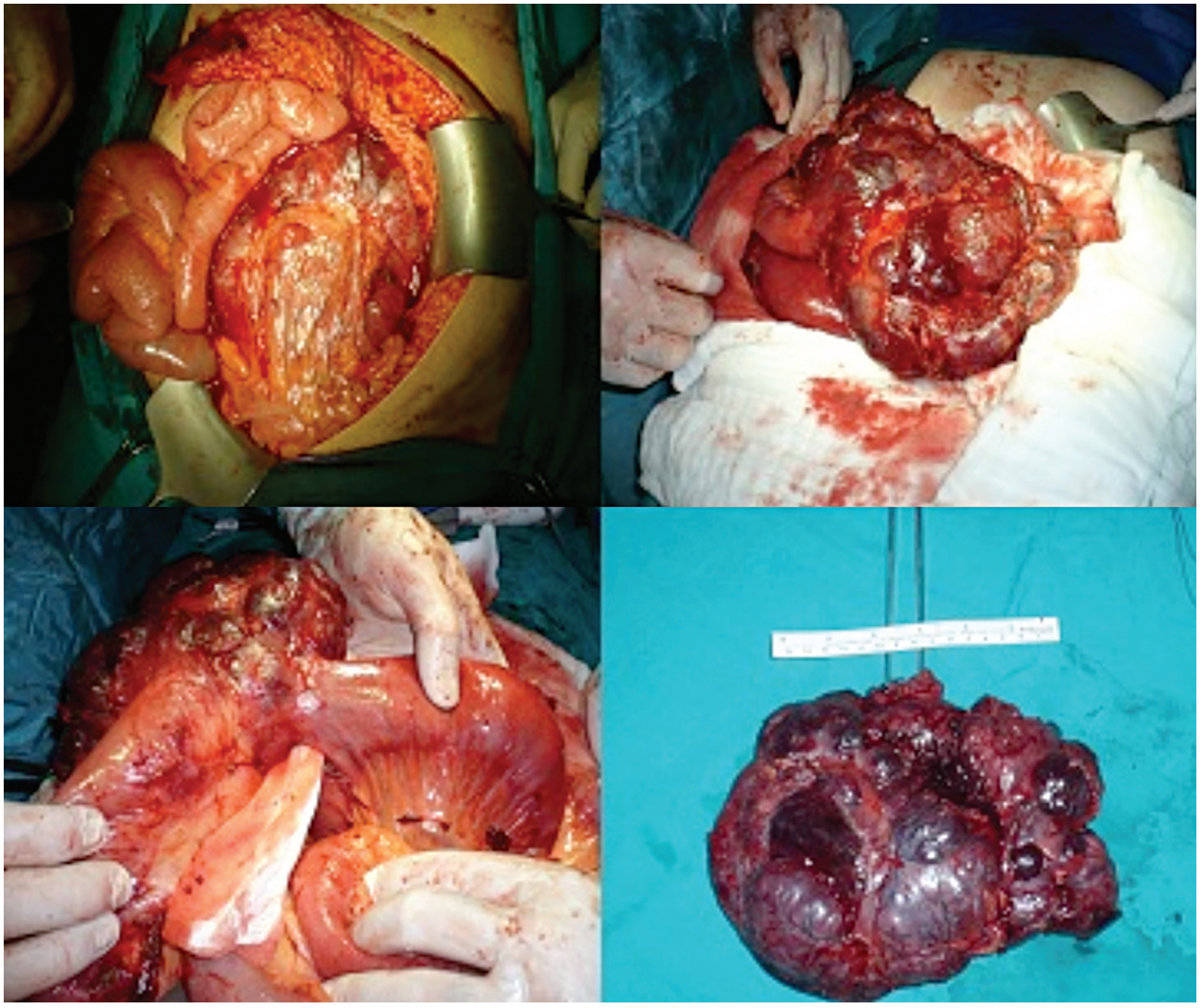

Figure 2. A mass measuring 20-cm in diameter extending from the anterior abdominal wall to the left retroperitoneal space.

tected in the solid organs. The mass was completely excised with the involved small intestinal segment, and end-to-end bowel anastomosis was performed. Intraoperative frozen section of the excised specimen was performed, and a diagnosis of mesenchymal tumor was made. Postoperative histopathologic examination revealed a high-grade small intestinal GIST (Figures 3, 4). Immunohistochemically, the tumor was positive for C-KIT (100%), smooth muscle actin (SMA, 100%) and S100 (<5%), and negative for desmin and CD34.

Following an uneventful postoperative course of five days, the patient experienced symptoms and signs of pulmonary emboli. Upon radiologic confirmation of the emboli diagnosis, the patient received low molecular weight heparin treatment (enoxaparin 0.8 cc) and was discharged from the hospital on the 24th day with no other postoperative complications. After discharge, the patient was treated with adjuvant chemotherapy with imatinib. No recurrence of the disease has been detected over a follow-up period of one year.

DISCUSSION

GISTs are the most common mesenchymal tumors of the GI tract. The etiology and pathogenesis of these tumors are unknown. Their incidence increases between 60 to 70 years of age and they are predominant in males (3).

GISTs smaller than 2 cm in diameter are generally asymptomatic and are detected incidentally during barium studies, endoscopy or abdominal scans performed for other reasons. The most common signs and symptoms of GISTs include abdo-

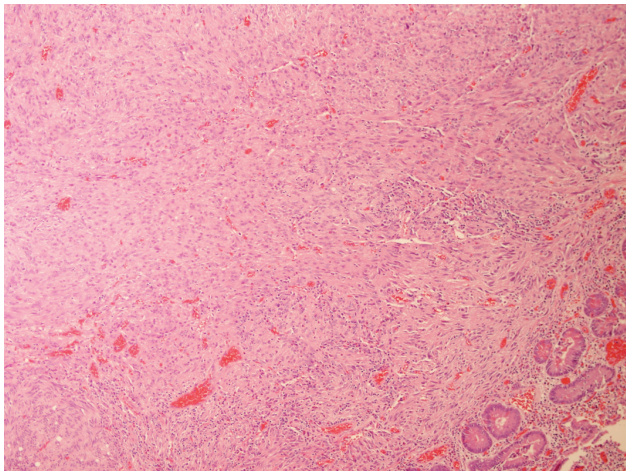

Figure 3. Diffuse tumoral infiltration throughout the intestinal wall reaching the mucosal surface (hematoxylin & eosin, x100).

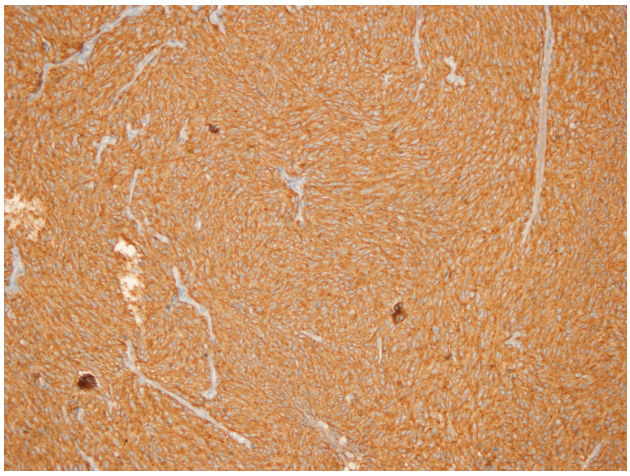

Figure 4. Tumoral cells show strong cytoplasmic CD-117 immunoexpression (CD-117, C-KIT, x200).

minal pain, abdominal mass, GI hemorrhage, partial or complete small bowel obstruction, weight loss, and fever or abscess (4). Karagulle et al. (5) reported a rare case of multifocal GISTs of the small intestine presenting with an abscess secondary to intestinal perforation. Crosby et al. (4) also reported that abdominal pain and abdominal mass are the most common presenting symptom and sign of GISTs. In the presented case, the patient's symptoms and signs were consistent with these earlier reports.

For the diagnosis of GISTs, several radiologic modalities such as CT, barium studies and/or angiography are used; however, none of these modalities can establish the correct diagnosis with 100% certainty. Preoperative fine needle aspiration biopsy is not indicated because of the risk of tumor rupture and intraperitoneal seeding (1).

The mechanism of bowel perforation secondary to GIST is unclear. Possible suggested mechanisms include increased intraluminal pressure due to tumor obstruction, replacement of the bowel wall by tumor cells followed by necrosis, and bowel ischemia due to tumor embolization (6). In the presented case, the necrosis in the tumor may be the reason for the closed perforation.

GISTs typically express CD117 (KIT), often CD34, and sometimes SMA. Desmin is seen only occasionally. S100 positivity is rare, but is seen most frequently in the small intestinal GISTs. In the presented case, C-KIT, SMA and S100 were positive, whereas CD34 and desmin were negative. Since the tumor was negative for CD34 and positive for S100, it was concluded to have originated from the small bowel.

The treatment of choice is surgical excision of the tumor and resection of all the infiltrated tissues, if any. However, lymph node dissection is not recommended by many authors (6,7). Conventional chemotherapy has a poor effect on GISTs. Radiotherapy can be used for the treatment of GISTs only for analgesic purpose and for intraabdominal hemorrhage, if the accurate location of the tumor is known (1,7,9).

For metastatic and non-resectable GISTs, the substance STI-571 (imatinib), which is a selective inhibitor of tyrosine kinases, platelet-derived growth factor receptors and C-KIT receptors can be used. Sunitinib is a small molecule that inhibits multiple receptor tyrosine kinases (10), and has a similar mechanism of action as imatinib. In the presented case, although there was no metastasis, the patient was prescribed imatinib for three months due to the closed bowel perforation.

In conclusion, small intestinal GISTs can present with vague symptoms and signs; therefore, an exact diagnosis preoperatively is generally difficult. It must be kept in mind that, although rare, GISTs can also present with closed bowel perforation, as seen in the presented case.

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